Modern Concepts of Cardiovascular Disease

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SCLEROSIS OF THE PULMONARY ARTERY

Etiology and Pathogenesis

In 1893 Huchard compiled data indicating the relative infrequency of sclerosis of the pulmonary artery as compared with those of the systemic circuit. It is by no means a rare condition. The circumstances under which it occurs indicate three major possibilities as causative agents in its production; (1) pulmonary hypertension, (2) pulmonary arteritis, and (3) anoxemia. It is at once apparent that anoxemia is likely to be a common factor with either of the other two.

The commonest condition with which it is associated is mitral stenosis. It is also found in all forms of congenital heart disease in which the pressure in the lesser circuit is heightened by abnormal communications with the greater. It is at once apparent that anoxemia is a common factor with hypertension in both groups of these disorders.

Sclerosis of the pulmonary arteries is also associated with a wide variety of types of fibrosis of the lungs. It occurs in the advanced pneumonoconioses, with the infectious fibroses of all types, tuberculous, mycotic, and chronic non-tuberculous respiratory infections. In this group of cases it has been commonly assumed that the fibrosis caused an increased resistance in the pulmonary circuit. However, it is readily apparent that anoxemia may be an equally important factor in this group as well, in fact it may be the sole factor, because the existence of pulmonary hypertension in the fibroses is purely an assumption, since direct measurement of the pulmonary blood pressure in man is impossible.

Arteriosclerosis of the lesser circuit also occurs in severe cases of emphysema. It has generally been assumed that emphysema causes an increased resistance to perfusion of the lung. However, recent work of Alexander, Luten and Kountz has shown that the elevated intrapleural pressure in emphysema tends to impede the venous return to the thorax so that the increase in peripheral resistance in emphysema may be offset by a diminished output of the right ventricle. Anoxemia is a constant and demonstrable factor in severe emphysema.

Various forms of arteritis have been observed in the pulmonary circuit. Of these, syphilis appears to be important. In some cases a syphilitic process may be observed in the first portion of the artery identical in appearance with that of the aorta, and usually associated with an aortitis. In other cases, such as that of Konstam and Turnbull, a wide spread syphilitic arteritis and pneumonitis is observed, to which the term Ayerza's disease had been applied by his pupils, Arrillaga and Escudero. In such cases atheromatous as well as inflammatory change may occur.

An interesting case was reported by Mocaigne and Nicaud, of an infection with *aspergillus funigatus,* which caused an extensive and thrombosing pulmonary arteritis and atherosclerosis together with a fibrosis of the lung. Rheumatic arteritis has been recently described by Von Glahn and Pappenheimer. In view of the close association of pulmonary sclerosis with mitral stenosis, one must naturally consider the possibility of a relationship between rheumatic pulmonary arteritis and the subsequent sclerosis. The evidence so far does not favor the existence of any direct relationship between them. The healing stages of the arteritis are well known and are not such as would be confused with atherosclerosis.

The possibility of a primary pulmonary sclerosis has been frequently considered since Romberg in 1891 published a case apparently of this type. Leonard Rogers described 10 cases of pulmonary sclerosis among Bengalis ranging in age from 12 to 55, in which there was neither mitral nor congenital heart lesions, nor general arteriosclerosis. The descriptions of the lungs of his cases are meagre or entirely lacking in the report.

Clarke, Coombs, Hadfield and Todd report two cases in children of pulmonary arteriosclerosis without obvious cause. In another child a dilatation of the pulmonary artery without sclerosis was observed in association with congenital abnormalities in the lung (bronchiectases cysts?). The writer has recently observed exquisite pulmonary arteri-

osclerosis in a man of 35 years, who died as a result of two large cysts in each lung, which were thought by some to be emphysematous and by others to be congenital cysts. There was considerable fibrosis in the portions of the lungs not involved by the cysts.

As one reviews case reports of the conditions with which pulmonary sclerosis is associated, it is difficult to find any single factor as constant as that of anoxemia. In support of the view that this may be the essential factor in the production of the pulmonary sclerosis, is the observation of Campbell, who produced this lesion in cats by exposing them for 33 days in a low pressure chamber. The anatomical change in the vessels was described as marked hypertrophy of the *tunica muscularis*. The animals exhibited the passive congestion of viscera characteristic of congestive heart failure.

Hurtado in his studies of the natives of the high Peruvian Andes observed the frequent occurrence of pulmonary sclerosis at autopsy in association with a chronically hyperaemic state of the lungs, which has subsequently been shown to occur at low barometric pressures. (Hurtado, Kaltreider and McCann, 1934).

In summing up the evidence regarding pathogenic mechanisms in the production of pulmonary sclerosis the two important factors appear to be *anoxemia* and *hypertension*. Arteritis appears to have little direct effect, except in so far as vascular occlusion and accompanying fibrosis contribute to anoxemia and hypertension in the pulmonary circuit.

Symptoms

There are probably few symptoms directly referable to the arterial disease, apart from those of the primary condition producing it. Many writers refer to a peculiar *angina hypercyanotica* in association with pulmonary sclerosis with the inference that the pain may arise in the pulmonary artery. This is purely speculative. The anoxemia indicated by the extreme cyanosis could activate the ordinary mechanisms by which angina pectoris is mediated.

The syndrome of Ayerza describes fairly well the terminal state of most conditions in which pulmonary sclerosis occurs. A marked degree of cyanosis, frequently with polycythemia and clubbing of the fingers is observed giving rise to the name "cardiacos negros," the "black cardiacs." The dyspnea accompanying it is frequently not associated with orthopnea. Cough and mucopurulent sputum with heart failure cells are usual, and hemoptyses are not infrequent. The patients are torpid, drowsy and somnolent. They tolerate morphia poorly and frequently die during sleep. Edema, peripheral venous engorgement, enlargement of the liver and ascites give evidence of the dilatation of the right heart, which can be confirmed by percussion, and by roentgenographs of the chest. The enlargement of the pulmonary conus can frequently be demonstrated by percussion and palpation in the third left interspace over which a rough systolic murmur is sometimes heard, followed by an early diastolic whiff if the pulmonary valve is incompetent. The roentgenographic demonstration of the pulmonary artery is usually easy and highly characteristic.

The physical signs arising in the lungs depend upon the nature of the associated pulmonary lesion, and the degree of passive congestion. An electrocardiogram, highly characteristic of the Ayerza syndrome, is frequently observed. It gives evidence of deviation of the electrical axis to the right, and as myocardial failure becomes established inversion of the T waves in leads II and III is usual. These special characteristics may be lacking due to the presence of pre-existing disease on the left side of the heart.

Diagnosis

The presence of pulmonary sclerosis may be suspected whenever a high degree of cyanosis persists in any of the conditions with which it is associated, such as marked mitral stenosis, congenital heart disease, marked emphysema, any of the pronounced forms of pulmonary fibrosis, and certain congenital abnormalities of the lungs.

The diagnosis may be made with certainty when, in addition to the above, dilatation of the pulmonary artery and right sided enlargement are demonstrable by physical signs and roentgenographs and when the typical electrocardiogram of the *cor pulmonale* is observed, as described above. If the terminal picture of the Ayerza syndrome is one of considerable duration pulmonary sclerosis will almost invariably be found postmortem.

Treatment

In the management of patients with the Ayerza syndrome the administration of oxygen in a tent is frequently more effective than digitalis in combating heart failure, though digitalis should be exhibited in the usual manner. Morphia should be used in these cases with extreme caution. Venesection, and thoracentesis, for relief of hydrothorax, and sometimes the insertion of Southey's tubes are useful manoeuvers in the management of these cases.

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